# ABSTRACT

## CLINICAL PROFILE AND OUTCOME OF PRIMARY IMMUNODEFICIENCY IN CHILDREN AT A TERTIARY CARE CENTRE IN SOUTH INDIA

**OBJECTIVES**:

To study the frequency, clinical features, disease complications and outcome of primary immunodeficiency disorders in children.

**METHODOLOGY:**

Prospective observational study (December 2017 to September 2018) done at ICH&HC, Madras Medical College, Chennai.

STUDY POPULATION: Children in the study age group admitted in ICH, satisfying the inclusion criteria.

SAMLPLE SIZE: The study covered all the children during the study period who presented with complaints in the inclusion criteria.

PROCEDURE: All children who meet the inclusion criteria were included in the study and informed consent were obtained from the parents. Detailed history with anthropometric details, vital signs, clinical examination and detailed systemic examination were done. The necessary investigations to diagnose PID were done. Statistical analysis was done using SPSS software.

**RESULTS:**

Thirty children were diagnosed to have primary immunodeficiency. There were 22(73.3%) males and 8(26.7%) females. The minimum and maximum age of presentation was 2 months-10 years. 13 children had severe combined immunodeficiency(SCID), 5 children had chronic granulomatous disease(CGD), 4 children had common variable immunodeficiency(CVID), 3 children had Wiskott Aldrich Syndrome(WAS), 2 children had selective IgM deficiency, 2 children had Leukocyte Adhesion Defect and one child had Bruton's agammaglobulinemia. Common mode of presentation were pneumonia in all the children, failure to thrive in 23, otitis media in 11, recurrent diarrhea in 10, tuberculosis in 8, skin and soft tissue infections and candidiasis in 6 and 1 each of meningitis, osteomyelitis and lymphadenopathy. History of consanguinity was there in 13 children. Family history of sibling deaths was elicited in 3 families. 13 children (56.7%)

were died during the study period. 10 out of 13 children who died were SCID. Infectious agents were isolated in 12 cases.

**CONCLUSION:**

Based on the results of our study, we think that primary immunodeficiency diseases occur in our country more than we expect. We are lack in reporting the diagnosed cases of PID, so the exact prevalence of the disease is not known. Reporting these diseases, sharing data and experience about managing these rare disorders will make a major difference in the future.

**KEY WORDS:** Primary immunodeficiency, Severe combined immunodeficiency, chronic granulomatous disease, combined variable immunodeficiency